Congenital Atresia of the Esophagus and Tracheoesophageal Fistula

Review of 48 Cases

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• **Forty-eight cases of esophageal atresia and tracheoesophageal fistula observed at the University of California Medical Center, San Francisco, over a 25-year period (1937-1961) were reviewed. Four of the patients were born at the University of California Hospitals, a frequency of 1 in 6,823 births. In 43 of the 44 referred cases, the referring physician established the diagnosis before admission by passing a tube into the blind esophagus or by radiography.**

Survival rates are comparable to those of other medical centers and emphasize the importance of prompt diagnosis, skillful repair and meticulous preoperative and postoperative care.

THE PURPOSE of this paper is to present the experiences at the University of California Medical Center, San Francisco, in the management of congenital atresia of the esophagus and tracheoesophageal fistula.

The first patient with esophageal atresia and tracheoesophageal fistula on record at this center was born at the University of California in 1937. In the ensuing 25 years (through 1961), 48 patients with these anomalies were admitted to the University of California Medical Center. All diagnoses were confirmed at operation or autopsy. Four of the patients were born at the University of California Hospital (a ratio of 1 to 6,823 births); 44 were referred from Northern California physicians. Twenty-five were females and 23 males; 11 were premature (by weight—2,500 gm or less). In 47 cases the lesions were of Ladd type III,* and one had an "H" type fistula without esophageal atresia. In all cases but one (the "H" type fistula), the referring physicians had established the diagnosis before admission by passing a tube into the blind esophagus or by demonstrating the nature of the anomaly by radio-opaque contrast study.

The presenting symptoms during the first hours or days of life were excessive mucus, choking, cyanosis, sputtering and regurgitation of feedings.

The average birth weight of the 48 babies was 2,850 grams; that of the 12 survivors was 2,945 grams, and of those not surviving operation 2,766 grams. The infants surviving surgical repair were operated upon at an average age of 2.25 days; for those who died the average was 3.81 days. Average survival (in days) of the infants who underwent operation is compared with survival time of those treated medically in Table 1. Causes of death in 36 cases are listed in Table 2. Table 3 shows the types of anomalies found in 18 of the 34 infants at autopsy. In five of these babies, the associated anomaly was the probable cause of death or contributed to it.

TABLE 1.—*Nature of Treatment and Length of Survival of 48 Patients with Esophageal Atresia and Tracheoesophageal Fistula*

No. Cases	Condition	Average Days Survival
7	No operation	10
2	Fistula ligation only	3½
2	Gastrostomy only	11½
1	H-type operation	7½ mo.
1	Stillbirth	....
12	Ladd III ligation and anastomosis	living
23	Ladd III ligation and anastomosis	21

TABLE 2.—*Causes of Death in 36 Cases of Esophageal Atresia and Tracheoesophageal Fistula*

	No. Cases
Without operation—	
Aspiration pneumonia and atelectasis	5
Brain hemorrhage	1
Malnutrition and dehydration	1
	7
Stillbirth	1
Operation—	
Mediastinal hemorrhage or infarction, hydrothorax, emphysema, pulmonary edema, atelectasis (postoperative); in 5 of the 11 the anastomosis had dehisced	11
Aspiration pneumonia (due to refistulization)	5
Aspiration pneumonia (dating prior to surgery)	5
Congenital heart disease	3
No autopsy	2
Asphyxia (post-tracheostomy)	1
Malnutrition and dehydration (diarrhea)	1
	28

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*Ladd type III, the most common variation, consists of esophageal atresia with a blind upper pouch and a lower esophageal segment which communicates at its lower end with the stomach and at the upper end with the trachea, the latter through a fistulous tract at the region of the tracheal bifurcation.

TABLE 3.—*Congenital Anomalies Associated with Esophageal Atresia and Tracheoesophageal Fistula in 18 Cases*

Gastrointestinal	11	(Meckel's diverticulum, 5; accessory spleen, 3; ectopic pancreas, 3; imperforate anus, 2; cloacal anus, absent gallbladder, pyloric stenosis, gastric hernia and lymphangiectasis of colon, 1 each)
Cardiovascular	8	(Patent ductus arteriosus, 5; coarctation of aorta, 3; aberrant right subclavian artery, 2; atrial septal defect, absent left umbilical and iliac arteries, bicuspid pulmonary and aortic valves, 1 each)
Genitourinary	7	(Fused kidneys, 2; absent kidneys, hypoplastic kidneys, anomalous ureters, hypospadias, bicornate uterus, double ureters, 1 each)
Other anomalies	8	(Hemivertebrae, 2; microgyric occipital lobes, cavernous hemangioma of choroid plexus, cleft palate, absent right thumb and hypoplastic left thumb, double rib, 1 each)

DISCUSSION

Esophageal atresia and tracheoesophageal fistula is a relatively uncommon anomaly. The incidence at UC Medical Center, San Francisco, was 1 in 6,823 births in the 25-year period reviewed; other investigators[13] have estimated its incidence as 1 in 2,500 births. This combination of anomalies was first described by Gibson[5] in 1703. Leven[10] and Ladd[9] contributed to recent interest by their surgical successes in 1939.

Early diagnosis, skillful surgical repair and meticulous preoperative and postoperative management continue to influence survival rates, which have ranged in various series from 36 to 62 per cent.* In the present series (up to 1953) five of the 22 infants (23 per cent) undergoing ligation of the fistula and anastomosis of the esophageal atresia survived. Since 1953, seven of thirteen (54 per cent) have survived. Other factors that appear to have prevented a higher survival in this series were dehydration, malnutrition, prematurity, aspiration pneumonia (in many cases present before operation), the presence of co-existing anomalies—especially cardiovascular—and an unusually long atretic esophageal segment.

*Reference Nos. 1-4, 6-8, 11, 12, 14.

The "H" type fistula presents a special problem in both diagnosis and treatment. Diagnosis in the one case in the present series was dramatic and accidental—a nurse noted bubbles emerging from the tip of a feeding tube which had been passed half way down the esophagus. Surgical repair was technically successful but the child died.

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